Short-term Results of Kasai Operation for Biliary Atresia: Experience from One Institution

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BACKGROUND: The purpose of this study is to review the short-term outcome of patients with biliary atresia (BA) treated by the Kasai operation at our institution.
METHODS: Ninety-two BA patients treated by the Kasai operation between January 1996 and December 2002 were reviewed. The diagnosis of BA was confirmed by intraoperative cholangiography. The outcome of treatment was categorized into two groups: jaundice-free (total bilirubin < 2 mg%) and persistent jaundice (≥ 2 mg%). The outcome of Kasai operation was evaluated 1 year after surgery. Data are expressed as mean ± SD.
RESULTS: Average age at the time of surgery was 90.26 ± 36.44 days. Only 22.8% (21/92) of patients had Kasai operation before 60 days of age. Histologically, 49 patients (54.4%) had liver fibrosis at the time of surgery. Of 92 patients, 17 were not included in outcome evaluation as they were less than 1-year post-surgery. Therefore, 75 patients could be evaluated for the outcome. Thirty-eight patients (50.67%) were jaundice-free 1 year after surgery. Liver histology and age at the time of the Kasai operation did not influence early outcome. The most common complication was ascending cholangitis.
CONCLUSION: Half of our BA patients who underwent Kasai operation were jaundice-free 1 year after surgery. The lack of impact of age and liver pathology on outcome is presumably due to the briefness of the follow-up. In general, our patients underwent Kasai procedure too late. It is therefore important for us to conduct a campaign to highlight the plight of these patients and the urgency of referral for neonates with jaundice. [*Asian J Surg* 2006;29(3):188–92]

Key Words: biliary atresia, cholangitis, Kasai operation

Introduction

Biliary atresia (BA), a progressive inflammation and obliteration of both extra- and intrahepatic bile ducts, is a common problem in early infancy with cholestatic jaundice. The outcome is eventual death from liver decompensation or infection when they are left without any treatment, usually by the 2nd year of life.[1] The introduction of hepatic portoenterostomy by Kasai et al to correct obliterated extrahepatic bile ducts has improved patient survival.[2] Most surgeons attribute the successful use of the Kasai operation to early diagnosis, careful surgical procedure and prevention of postoperative cholangitis.[3–5] Nevertheless, liver disease, resulting from antenatal involvement of the intrahepatic bile duct, progresses in patients who are not treated. The progressive disease culminates in the end-state of cirrhosis for these patients who will eventually require liver transplantation.

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In order to define particular aspects of the treatment, which need to be emphasized, understanding of the therapeutic results is necessary. The purpose of this study is therefore to review the short-term outcome of patients with BA treated by the Kasai operation at our institution.

Materials and methods

During the period from January 1996 to December 2002, data of all consecutive infants with BA treated by hepatic portojejunostomy with Roux-en-Y (original Kasai operation) from the medical records at King Chulalongkorn Memorial Hospital were reviewed for the following data: gender, age at the time of surgery, pre- and postoperative liver function tests, associated malformations, operative findings and postoperative complications. The majority of BA patients were investigated for diagnosis by both ultrasonography and hepatic scintigraphy. The diagnosis of BA was confirmed by coeliotomy with intraoperative cholangiography before undergoing Kasai operation.

The outcomes of Kasai operation were assessed at 1 year after the procedure and categorized into two groups: jaundice-free (total bilirubin levels < 2 mg%) and persistent jaundice (total bilirubin levels ≥ 2 mg%). None of the patients in this study showed symptoms and signs of fever or ascending cholangitis (high fever, elevated serum total bilirubin and leukocytosis) at the time of outcome evaluation. Types of BA were classified as previously described by Nio and Ohi: type I, atresia of common bile duct; type II, atresia of common hepatic duct; type III, atresia at porta hepatis.[6] The association between outcome and age at the time of Kasai operation as well as liver pathology was analysed using χ^2 tests. All data are expressed as mean ± SD. For all statistical analyses, SPSS version 10.0 (SPSS Inc., Chicago, IL, USA) was used.

Results

Ninety-two BA patients underwent Kasai operation during the review period. There were 42 (45.7%) males and 50 (54.3%) females with a mean age at the time of surgery of 90.26 ± 36.44 days (range, 30–210 days). Only 22.8% (21/92) of the BA patients had the operation before the age of 60 days (Figure). Out of the 92 patients, 68 (73.9%) presented with clinical jaundice at birth while the remaining (26.1%) developed recognizable jaundice after the age of 1 month. Associated anomalies consisted of cardiac anomalies in three patients (tetralogy of Fallot, pulmonary stenosis and patent ductus arteriosus), Turner's syndrome in one patient, acquired prothrombin complex deficiency in one patient, glucose-6-phosphate dehydrogenase enzyme deficiency in one patient and ectopic pancreas at jejunum in one patient. There was no polysplenia syndrome detected in this series.

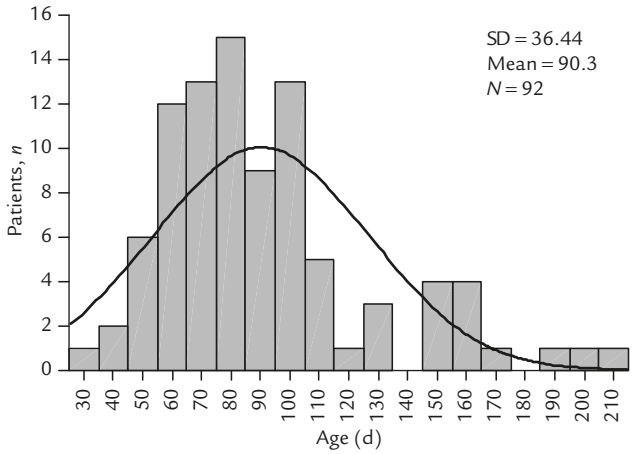

Figure. Age of patients at the time of Kasai operation.

Among the 72 patients investigated by abdominal ultrasonography, 51.4% (37/72) had visualized gallbladder reported by radiologists. Eighty-six patients had been investigated by hepatobiliary scintigraphy prior to operation, and all 86 showed no radiotracer in the intestine during the 24 hours following scintigraphy. Mean preoperative liver function levels were as follows: total bilirubin, 12.31 ± 5.58 mg%; direct bilirubin, 9.12 ± 4.05 mg%; alkaline phosphatase, 1,009.67 ± 522.64 IU/mL; serum glutamic oxaloacetic transaminase, 262.27 ± 150.01 IU/mL; serum glutamic pyruvic transaminase, 174.08 ± 109.79 IU/mL.

As a result of intraoperative cholangiography, 94.6% (87/92) of patients had BA type III and the remaining five had BA type I. There was no BA type II detected in our series. All patients with BA type III, 44.8% (39/87), had patent common bile duct while the remaining had atresia of the common bile duct as well as porta hepatis. Mean operative time was 206.8 ± 40.6 minutes (range, 105–315 minutes). Pathological report of liver biopsy revealed that 54.4% (49/90) already had liver fibrosis at the time of operation.

Perioperative complications occurred in eight patients (8/92 or 8.7%). The complications were prolonged bile leakage in three patients, wound infection in two patients,

sepsis in two patients (both of which resulted in death) and intestinal perforation at the site other than anastomosis in one. Postoperative complications comprised cholangitis in 34 patients (38%), gastrointestinal bleeding from varices in nine patients whose livers were cirrhotic at the time of Kasai operation (10%) and intestinal obstruction in two patients (2%). Among 34 patients with cholangitis, 21 (61.76%) had a single episode only and the remaining 13 (38.24%) had recurrent episodes.

Among the 92 patients, 17 patients were less than 1-year post-surgery; therefore, only 75 patients could be evaluated for postoperative outcomes. Of these, 38 (50.67%) were jaundice-free, and 37 (49.33%) had persistent jaundice. In the good outcome group, the mean duration between the operation and jaundice-free onset was 4.82 ± 3.86 months (range, 1–8 months). By using χ^2 tests, there was no association between outcome and either age or liver pathology at the time of the Kasai operation, as shown in Table 1.

Discussion

At present, the Kasai operation remains the most common initial management for BA patients at our institution.

Table 1. Outcome of Kasai operation and patient age and liver histology at the time of surgery

	Good outcome	Poor outcome	*p*
Age at operation			0.801
< 60 d	7	6	
≥ 60 d	31	31	
Liver histology			0.902
No liver fibrosis	18	17	
Liver fibrosis +ve	20	20	

In addition, the timing of surgical intervention for BA patients is a crucial factor. Most of the major series have confirmed that the best results from Kasai operation are achieved when patients have the surgery before the age of 60 days.[7–12] An analysis of the BA patients treated by the Kasai operation at our institution showed that most of them (77.2%) had the surgery quite late. Only 22.8% had the Kasai operation before 60 days of age. Mass screening for BA patients by examining all neonatal stools with stool colour cards, which was previously proposed by Maki et al,[13] may help patients with quicker diagnosis and surgery as a promising offensive policy. Alternatively, a campaign towards primary healthcare providers regarding the importance of early treatment needs to be urgently conducted.

Notwithstanding the late timing of surgery, 50.67% of our patients were jaundice-free at the 1-year follow-up. Previously published data extracted from 1985 to 1994 by Chandrakamol et al showed a jaundice-free rate of 35.7%.[14] The results indicate that there has been an improvement in surgical outcome at our institution. This may be attributed to our improved surgical techniques (mobilization of the liver and magnification using a surgical loupe during dissection of the porta hepatis) and more advanced knowledge in prevention of postoperative cholangitis (use of long-term oral antibiotics and short-term high-dose steroid administration). The jaundice-free survival rates reported by various authors range from 15.5% to 54.9%.[4,5,15–18] A large series reported by Kasai et al demonstrated that 84 of 245 BA patients (34.3%) achieved jaundice-free status after surgery (Table 2).[5] However, the outcome and the mean follow-up duration of BA patients in each institution were different; therefore, it is not appropriate to compare the results from different studies. The outcome of Kasai operation in this study was analysed at 1 year after surgery, which is quite short, and long-term follow-up has to be further evaluated.

Table 2. Jaundice-free rate in biliary atresia patients after Kasai operation

Reference	Period	Place	Patients, *n*	Jaundice-free rate, *n* (%)	Mean follow-up time (range)
Kasai et al[5]	1953–1987	Japan	245	84 (34.3)	1–34 yr
Lilly et al[15]	1973–1988	Colorado	125	35 (28.0)	86 mo (1–15 yr)
Lin et al[16]	1976–1989	Taiwan	60	23 (38.3)	7.2 yr (1–14 yr)
Wildhaber et al[29]	1974–2001	Michigan	81	31 (38.0)	92 mo (1–27 yr)
Carceller et al[30]	1974–1998	Quebec	63	27 (43.0)	1–28 yr
Current study	1996–2002	Thailand	75	38 (50.6)	1 yr after surgery

Approximately half of the BA patients (54.4%) had liver fibrosis at the time of surgery. Hays and Kimura[19] reported a relationship between liver fibrosis and prognosis. In their series, no infant whose liver biopsy showed severe fibrosis survived. Nevertheless, our data unfolded no association between outcome and the presence of liver fibrosis during surgery ($p > 0.05$). In addition, there was no association between outcome and the age of patients at the time of surgery. These observations are probably due to the short follow-up period. Longer period to assess outcome may reveal significant associations.

Ascending cholangitis is the most common postoperative complication, which was noted in 38% (34/90) of our patients after Kasai operation in addition to the common perioperative complication of abdominal surgery. Cholangitis frequently results in the cessation of bile flow, and repeated attacks cause a progressive deterioration in hepatic function. Prevention of cholangitis is therefore an essential factor for maintaining bile drainage. As a result, various types of antireflux procedures have been developed,[20–22] but none is superior to the original Roux-en-Y procedure.[23] At our institution, only hepatic portojejunostomy with Roux-en-Y or "the original Kasai operation" has been carried out since 1985 without additional modifications of the procedure. The incidence of ascending cholangitis in our series is comparable to the 40–60% rate of others.[3,24,25] Cholangitis is reported to be a risk factor associated with portal hypertension.[26] Because portal hypertension could develop even in BA patients who are jaundice-free, examination of oesophageal varices should be included in routine surveillance.

Despite the successful correction of extrahepatic biliary obliteration by Kasai operation, the on-going inflammation of intrahepatic involvement still exists. The on-going process elucidates why ultimate liver fibrosis is inevitable in a number of patients, who eventually require liver transplantation. Some authors have also improved survival rates via liver transplantation as a primary therapy for BA patients.[27,28] However, we think that liver transplantation should not be the first line of treatment in our country because of the shortage of liver donors and the very high cost of the operation. Therefore, liver transplantation in BA patients at our hospital was performed only in selected patients, who experienced end-stage liver cirrhosis and can comply with strict long-term follow-up.

In conclusion, half of our BA patients, who underwent Kasai operation, were jaundice-free at 1 year after surgery. The lack of impact of age and liver pathology on outcome is presumably due to the briefness of the follow-up. In general, our patients underwent the Kasai procedure too late. It is therefore important for us to conduct a campaign to highlight the plight of these patients and the urgency of referral for neonates with jaundice.

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